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Case 3

A 67-year-old man with a right thigh mass.

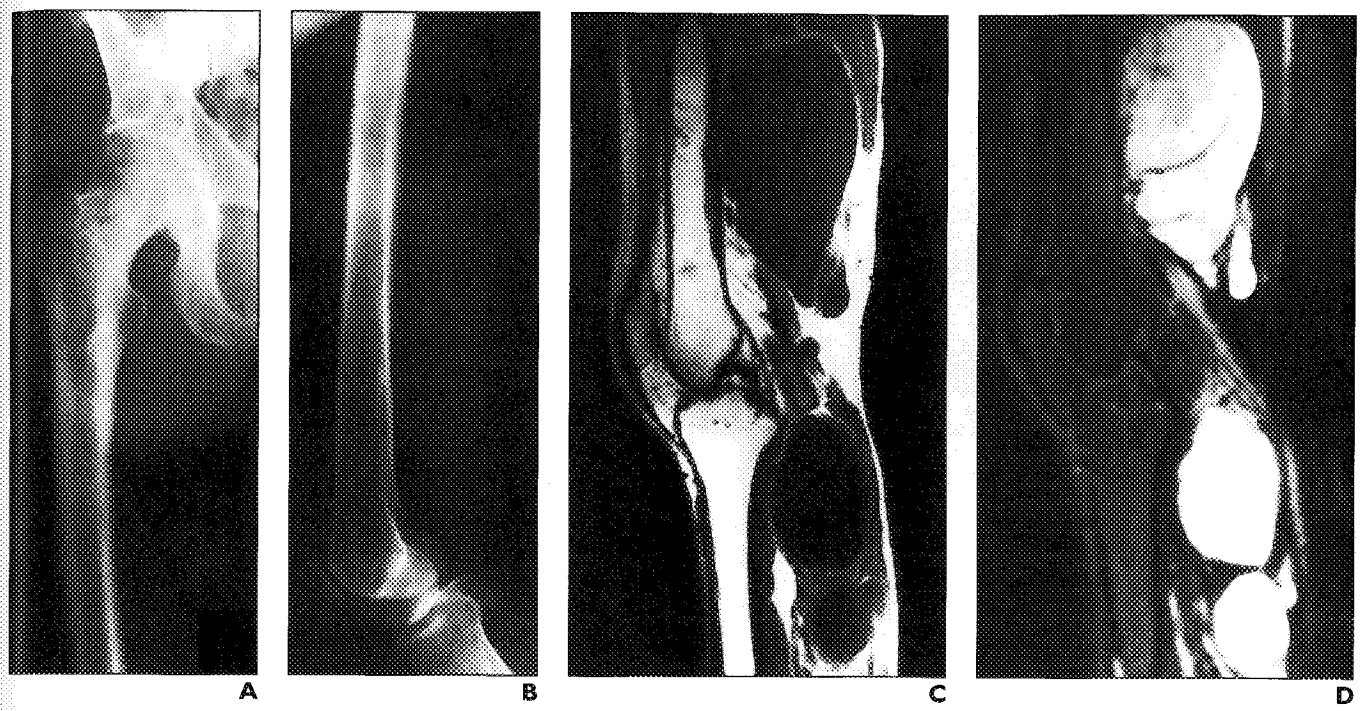

Fig. 3.—67-year-old man with right thigh mass.
A, Anteroposterior radiograph of right femur.
B, Lateral radiograph of right femur.
C, Sagittal spin-echo T1-weighted MR image of right thigh.
D, Sagittal fast spin-echo T2-weighted MR image with fat saturation of right thigh.

Case 2: Multiple Myeloma

Multiple myeloma is a common primary malignancy of plasma cells that occurs twice as commonly in adult men as in adult women. Myeloma accounts for 1% of all malignant conditions and 10% of hematologic malignant conditions [1]. Patients with myeloma often present with bone pain and fractures, anemia, impairment of renal function, or hypercalcemia. One of the most devastating complications is skeletal disease, which occurs in 80% of patients with multiple myeloma and is associated with extreme bone pain [2]. Diffuse and focal forms occur, and manifestations include generalized osteoporosis, expansile osteolytic lesions, and soft-tissue masses adjacent to areas of bone destruction (Fig. 2); pathologic fractures often follow. A sclerotic form of myelomatosis is rare, occurring in 1–3% of patients; sclerosis can also occur after chemotherapy or radiotherapy.

All existing methods of detecting skeletal myelomatous deposits have limitations. Bone lesions only become apparent on radiographs after the loss of more than 50% of bone mineral content at the site of the disease. Bone scans may have negative findings even in advanced myeloma, and CT has limitations in detecting marrow-based disease. Whole-body MR imaging is being established as an effective technique in oncologic screening [3]; it also has the potential to aid in the imaging diagnosis of myeloma and in directing a biopsy to the optimal area for pathologic yield.

Whole-body MR imaging is a sensitive, noninvasive method of examining a large proportion of bone marrow and the soft tissues of the skeleton. Because the proportion of fatty marrow in the spine increases with age, fat-suppression techniques offer important potential advantages for the depiction of bone marrow lesions on MR images. Short inversion time inversion recovery (STIR) sequences offer the advantages of high contrast between the normal and diseased tissues and suppression of the fat signal [4]. Diagnosis of bony lesions appears to be best achieved by fat-suppressed or T2-weighted imaging. MR imaging manifestations of myelomatous involvement include diffuse and focal hyperintense areas on T2-weighted imaging [5].

Fat-suppression techniques thus offer important potential advantages for the depiction of bone marrow lesions on MR images [6]. STIR imaging nulls the signal intensity of the fatty marrow; areas of high signal intensity reflect cellular marrow. Myelomatous infiltration can produce areas of T2 hyperintensity. Whole-body MR imaging may prove useful in showing the extent and complications of myeloma, as in this patient.

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Case 3: Mazabraud's Syndrome (Intramuscular Myxomas Associated with Fibrous Dysplasia of Bone)

Heneschen first described the association of fibrous dysplasia of bone and intramuscular myxomas in 1926 when he reported a case of multiple myxomatous tumors in a patient with osteitis fibrosa [1]. This rare association has since been sporadically reported in the medical literature, including case reports by Krogius in 1928 and Mazabraud and Girard in 1957 [2, 3]. In 1967, on reviewing the literature, Mazabraud et al. [4] proposed the existence of a syndrome characterized by the association of intramuscular myxomas and osseous fibrous dysplasia, now referred to as Mazabraud's syndrome.

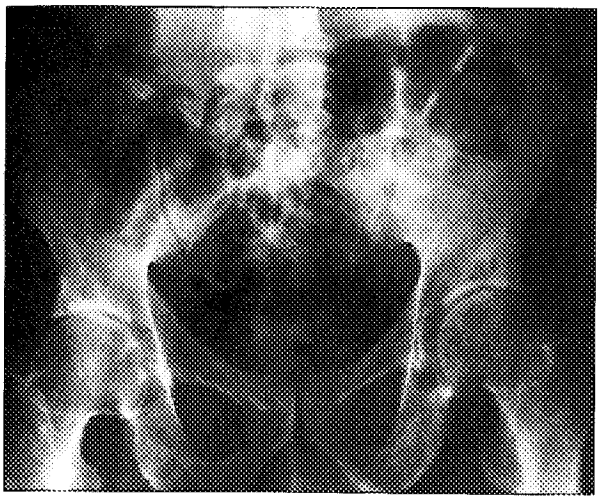
Osseous fibrous dysplasia is an uncommon benign tumor of unknown etiology characterized by fibroosseous metaplasia of one (monostotic) or multiple (polyostotic) bones. The monostotic form of the disease is much more common than its counterpart. Although any bone may be affected, the long bones, ribs, and skull are most often involved. Rarely, patients show extraskeletal manifestations of the disease, including pigmented cutaneous lesions (café au lait spots) and endocrine abnormalities. Complications include pathologic fracture and, in fewer than 1% of cases, malignant transformation, most commonly to osteosarcoma [5].

The diagnosis of fibrous dysplasia is usually made on radiographs, and advanced imaging is rarely necessary. The lesions are typically well defined and lytic, with a hazy or ground-glass matrix that is centered in the medullary cavity of the diaphysis of a long bone, most commonly the femur (Fig. 3). There may be expansile remodeling with associated endosteal scalloping, and the lesion may have a sclerotic border or rind. On skeletal scintigraphy, fibrous dysplasia usually exhibits increased activity, a finding that can be helpful to establish polyostotic disease. The matrix in osseous fibrous dysplasia may have low signal intensity on T2-weighted MR images, although most cases show a high-signal-intensity matrix. A hypointense sclerotic border or endosteal scalloping may also be apparent [6].

Myxomas are uncommon benign neoplasms that can occur in a variety of locations, including the heart, bones, skin, subcutaneous and aponeurotic tissue, genitourinary tract, and skeletal muscle. Although of uncertain origin, current evidence favors the concept that myxomas originate from primitive mesenchymal cells that differentiate as fibroblasts but lose their capacity to produce collagen and instead produce excess amounts of hyaluronic acid [7].

Intramuscular myxomas are rare in patients younger than 20 years and most frequently occur in adults with a mean age of 50–55 years. Myxomas are slightly more common in females than in males, although to our knowledge this female predilection has not been documented in myxomas associated with fibrous dysplasia. Most intramuscular myxomas are solitary, painless masses that tend to grow slowly or are stable in size for long periods of time before suddenly enlarging. They tend to involve the muscles of the thigh, buttocks, upper arm, and shoulder; usually lack a true capsule; and histologically may infiltrate the involved muscle [7].

On CT, intramuscular myxomas appear as well-circumscribed, homogeneous, low-attenuation, nonenhancing masses. Compared with skeletal muscle, these masses have low signal intensity on T1-weighted images and high signal intensity on T2-weighted and STIR images. Court-Payen et al. [8] described mild, inhomogeneous T1 enhancement after the administration of IV gadopentetate dimeglumine. The sonographic features of intramuscular myxomas were reviewed by Fornage and Romsdahl [9], who characterized these lesions as hypoechoic masses with small, fluid-filled clefts and cystic spaces and absent intratumoral flow on color Doppler examination.



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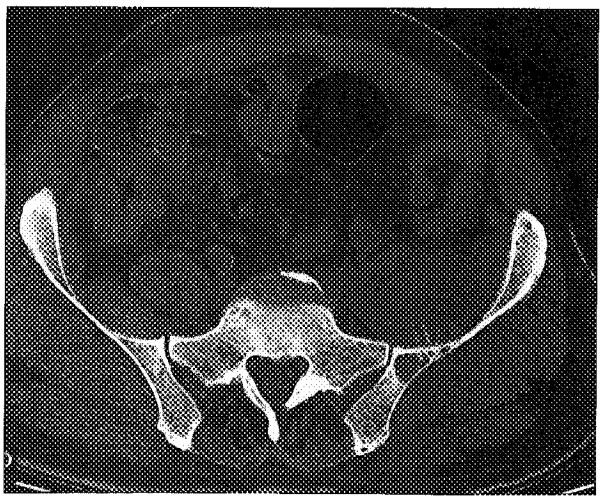


B

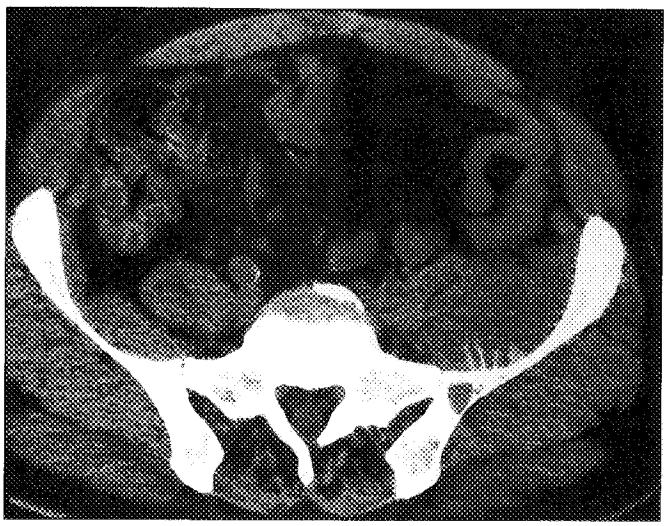
Fig. 2—66-year-old man with multiple myeloma.

A, Frontal radiograph of pelvis shows increased density over left ilium, raising suspicion of soft-tissue mass. Bilateral degenerative cystic disease of superior acetabula is incidentally noted.

B and **C**, CT scan of pelvis (bone windows) shows lytic lesion involving left ilium with sunburst spiculations extending into iliac muscle. Soft-tissue windows show enlargement of left iliac muscle resulting from plasmacytoma.

D, Sagittal T2-weighted MR image through thoracolumbar spine shows reduced height of T11 with posterior displacement of posterior portion of vertebral body with impression upon thecal sac.

E–G, Whole-body coronal turbo short inversion time inversion recovery MR images through lumbar vertebrae shows plasmacytoma involving left iliac muscle. Diffuse marrow hyperintensity resulting from diffuse myelomatous infiltration is seen. Focal signal hyperintensity within T11 vertebral body is caused by myeloma-induced compression fracture. High signal intensity in plasmacytoma in left iliac muscle is again seen.



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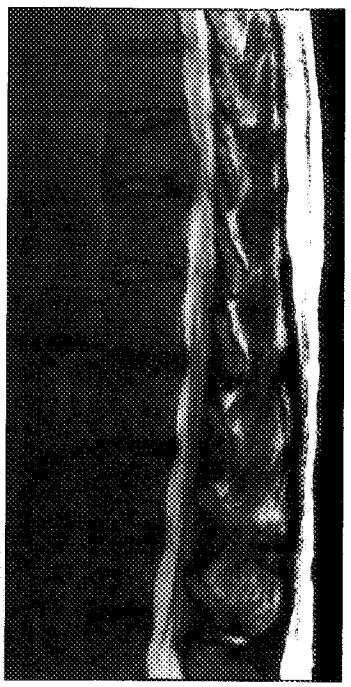

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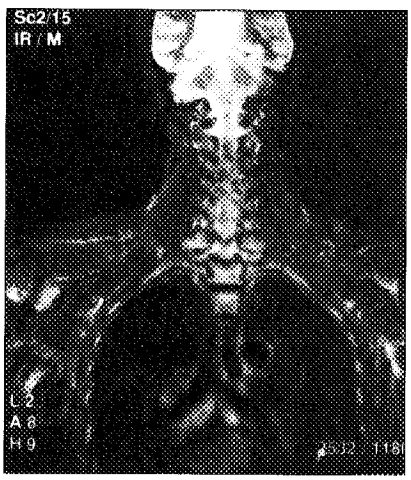

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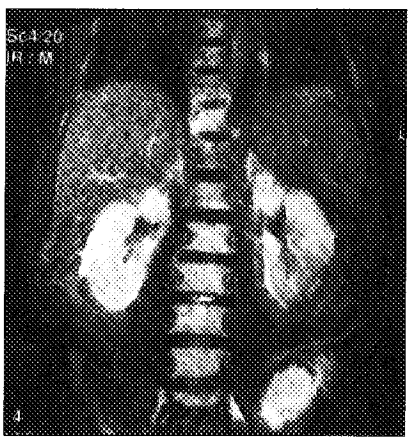

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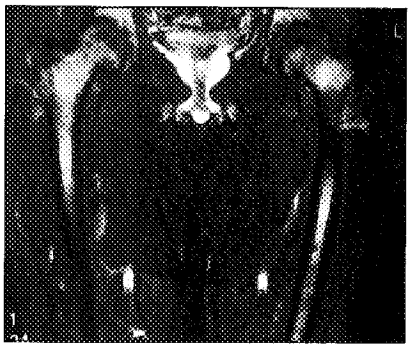

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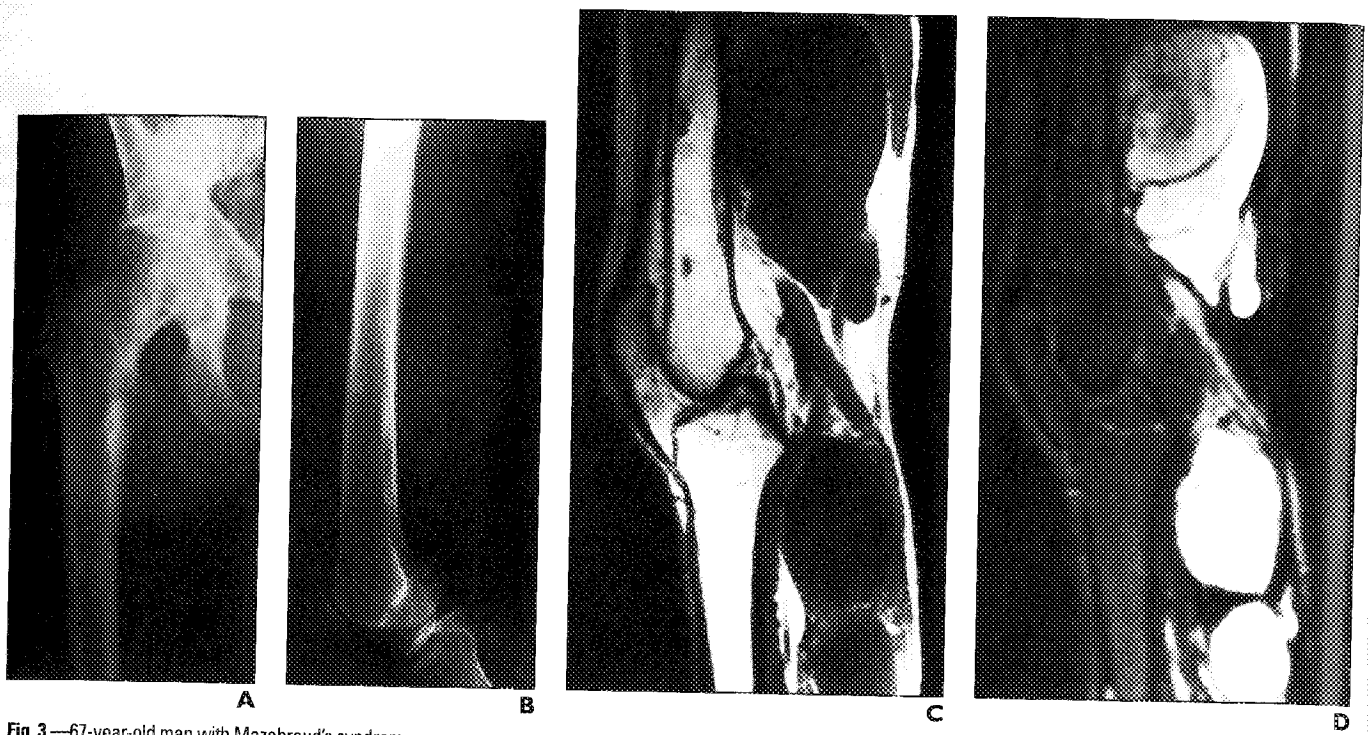

Fig. 3.—67-year-old man with Mazabraud's syndrome.
A and **B**, Anteroposterior (**A**) and lateral (**B**) radiographs of right femur show multiple soft-tissue masses without adjacent cortical erosion or periosteal reaction. Mixed areas of sclerosis and relative radiolucency in medullary cavity of proximal and distal femoral diaphyses with similar changes in intertrochanteric region of right femur are seen. Focal areas of cortical thickening and others areas with endosteal scalloping are also noted.
C, Sagittal spin-echo T1-weighted MR image of right thigh shows multiple lobulated, well-circumscribed, heterogeneous, predominantly low-signal-intensity soft-tissue masses in posterior aspect of thigh between femur anteriorly and hamstring muscles posteriorly. Displacement of adjacent musculature and neurovascular bundle without local invasion are seen. Additional masses were identified in leg along medial aspect of musculus vastus lateralis, in deep soft tissues of calf, and in gluteal muscles (not shown). Also seen are focal areas of decreased T1 signal identified in medullary cavity of distal femoral diaphysis and additional mixed regions of cortical thickening and endosteal scalloping. These masses showed mild, heterogeneous, predominantly peripheral enhancement on enhanced T1-weighted MR images with fat saturation (not shown).
D, Sagittal fast spin-echo T2-weighted MR image with fat saturation of right thigh shows masses to be predominantly high signal intensity. Although displaced, adjacent musculature is normal in signal intensity. No lesions were identified in left lower extremity (not shown). Note focal areas of mild T2 hyperintensity in medullary cavity corresponding to areas of decreased signal intensity on T1-weighted images.

In Mazabraud's syndrome, the intramuscular myxomas are often multiple and have a marked predilection for the right side of the body. As in the case we present, intramuscular myxomas tend to be located in the lower extremities, most commonly the thigh, and in the vicinity of the bones most severely affected by fibrous dysplasia. Although this syndrome is usually associated with the polyostotic form of fibrous dysplasia, its association with monostotic fibrous dysplasia, as in our case, has been previously described [7, 10]. Although the diagnoses of fibrous dysplasia and intramuscular myxomas were made simultaneously in the case we present, in most reported cases the diagnosis of fibrous dysplasia was made within the first three decades of life, preceding the diagnosis of myxomas by several decades. Occasionally, patients will exhibit skin pigmentation abnormalities with or without associated endocrinopathy—clinical findings not seen in our case.

Differentiating an intramuscular myxoma from other soft-tissue masses, such as a lipoma, a neurofibroma, or particularly a myxoid liposarcoma, can be difficult clinically, radiologically, and pathologically. Although certain imaging features may be helpful in narrowing the differential diagnosis, an excisional or core biopsy is often necessary to establish a definitive diagnosis. However, recognizing associated bone changes typical of fibrous dysplasia can aid in the correct preoperative diagnosis of an intramuscular myxoma and this rare, benign syndrome. Imaging can also be helpful in preoperative planning, specifically in establishing the relationship of the tumor to underlying neurovascular structures. Treatment depends on the extent of disease, but myxomas should be excised if the diagnosis is uncertain or if pain or pressure symptoms develop. When completely excised, the lesions rarely recur, although other myxomas may develop that were not detected at the time of initial assessment.

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